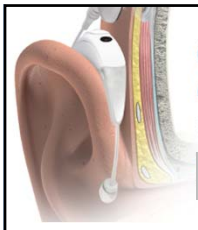


GRAND ROUNDS IN COCHLEAR IMPLANTS

Hosted by Advanced Bionics

Presented by Shelly Ash, M.S., CCC-A

September 25, 2015

Advanced Management of Complex Cases: Children with Multiple Medical Conditions

Advanced Management of Complex Cases: Children with Multiple Medical Diagnoses

Shelly Ash, MS, CCC-A
Audiologist and Cochlear Implant Team Coordinator
All Children's Hospital Johns Hopkins Medicine
St Petersburg Florida

Learner Objectives:

- The participant will be able to identify expansions in cochlear implant candidacy guidelines.
- The participant will be able to identify medically challenging conditions associated with hearing loss.
- The participant will be able to identify treatment options and prognoses for pediatric cochlear implant candidates and recipients with multiple medical diagnoses.

Introduction

- Cochlear implantation was introduced in the United States as a viable treatment for hearing loss in the 1980's
- Anticipated benefits were awareness of sound and enhancement of speech reading
- Early candidacy guidelines were very strict due to concerns that benefit would be limited or difficult to measure

Original FDA Guidelines
Pediatric Clinical Trials

- 24 months+ of age
- Profound hearing loss in both ears
- No benefit from traditional amplification
- No coexisting medical conditions
- Normal vision
- Normal intelligence
- No radiological contraindications
- Evidence of strong family support
- Appropriate expectations and motivation

Current FDA Guidelines
Pediatric Cochlear Implant Candidacy

- 12+ months of age
- 90 dB+ hearing loss in both ears 12-14 mos
- 70 dB+ hearing loss in both ears 24+ mos
- Marginal benefit from hearing aids
- No medical contraindications
- No radiological contraindications
- Strong family support
- Appropriate expectations and motivation

FDA Guidelines: Then and Now

- Age lowered from 24 to 12 months
  - Safety and efficacy data revealed cochlear implants were not dangerous
  - The primary goal of cochlear implants in children has shifted from awareness of sound to using audition to acquire language – earlier implantation is equated with closing the gap in language delay quicker

FDA Guidelines: Then and Now

- Currently infants under 2 years of age may be considered with a pure tone average of 90 dB HL or greater
- Children 2 years of age and older may be considered with a pure tone average of 70 dB HL or greater

FDA Guidelines: Then and Now

- "No benefit" vs "marginal benefit" from traditional amplification
  - Aided benefit originally was defined as audibility in the "speech banana"
  - Concerns regarding destroying residual hearing precluded candidates with audibility from being included
  - Current guidelines allow for pediatric candidates to have audibility in the "speech banana" as well as up to 30% open set understanding on word tests

FDA Guidelines: Then and Now

- Normal vision previously was required because cochlear implants were anticipated to assist with speech reading
- Current guidelines allow for visually impaired individuals to be considered, as performance data demonstrates benefit far beyond speech reading

FDA Guidelines: Then and Now

- Normal cognitive function was originally required as it was considered impossible to determine benefit if the candidate had a cognitive delay
- Presently it is widely accepted that individuals with less than normal intelligence can receive benefit from the cochlear implant

FDA Guidelines: Then and Now

- Persons with additional medical conditions were originally not implanted for many reasons
  - Risk of surgery vs anticipated benefit
  - Effect of chronic electrical stimulation on the brain
  - Concerns regarding post operative infection or other complications
  - Anticipated challenges in measuring benefit due to the impact of co-existing conditions
- Cochlear implantation is now recognized as a lower risk surgical procedure for the majority of individuals

FDA Guidelines: Then and Now

- Candidates whose radiological studies were atypical were not originally considered due to the possible limited insertion or derived benefit
- Currently candidates with ossification, fibrous tissue, cochlear malformations and enlarged vestibular aqueduct may be considered
- Lack of internal auditory canal, aplasic auditory nerve or absent cochlea are generally the only absolute rule out

FDA Guidelines: Then and Now

- Family Support and Appropriate Expectations and Motivation
  - Only unchanged guideline over time

FDA Guidelines: Then and Now

- Cosetti and Waltzman, 2012 reported factors that affect outcomes as well as influence broadening candidacy guidelines:
  - Advancements in technology
    - Improved speech understanding
    - Improved music appreciation
  - Improved surgical techniques and preservation of cochlear structures and residual hearing
  - The availability of electrophysiological measures
  - Internal device reliability

Children with Multiple Medical Diagnoses

- Why has the population of candidates with multiple medical diagnoses grown?
- The survival rate of premature infants increased substantially in the latter half of the 20th century
  - Low birth weight, oxygen dependency, delivery at <26 weeks gestational age carry greater risk for multiple medical diagnoses

Children with Multiple Medical Diagnoses

- Wakil et al, 2014 reported that 30 to 40% of children with sensorineural hearing loss also have additional developmental disabilities
- Hang et al, 2012 reported that 30% of sensorineural hearing loss is associated with a genetic syndrome
- Due to early intervention and early implantation, it is likely that an increasing number of children are implanted prior to certain disabilities or conditions being identified

Children with Multiple Medical Diagnoses

- Corrales and Oghalai, 2013, indicated:
- 25% of cases of congenital hearing loss are attributed to prenatal or postnatal disease or trauma
- 18% to undiagnosed genetic factors
- 15% to autosomal dominant genetic mutations
- 40% to autosomal recessive genetic mutations
- 2% to sex linked genetic mutations

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Verification of hearing loss
  - Use of behavioral and objective test measures
  - Parent questionnaire (IT MAIS)
  - Therapist and teacher observations

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Verification of hearing loss
  - Use of behavioral and objective test measures
  - Parent questionnaire (IT MAIS)
  - Therapist and teacher observations

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Evaluating this population requires a lot from the family
  - Parents/caregivers as informants
  - Parents/caregivers as the bridge between professionals
  - Parents/caregivers as decision makers

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Parents/caregivers as informants
  - Need to provide thorough and accurate information regarding the child's conditions
  - Need to give full disclosure regarding the child's conditions, therapies

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Parents/caregivers as the bridge between professionals
  - Discuss cochlear implant with other treating professionals
  - Provide information on cochlear implant to treating professionals
  - Request and encourage treating professionals to communicate with one another

Children with Multiple Medical Diagnoses: Cochlear Implant Candidacy Considerations

- Parents and caregivers as decision makers
  - Parents and caregivers should be empowered to weigh in and decide about participating in the evaluation process
    - Vaccinations
    - Anesthesia
    - Long term need for MRI
    - Sensory issues/tolerance of stimulation or equipment

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Verification of lack of benefit from traditional amplification
  - Evaluation measures should be selected based on chronological age, cognitive status and language age

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Medical Evaluation
  - Child must be healthy enough to undergo anesthesia
  - Child must be healthy enough to recover from the procedure with minimal risk of additional complications

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Radiological Studies
  - Verification of cochlear status and presence of an intact cochlear nerve
  - Identify any findings that may result in prolonging or complicating the procedure
  - Determine if one ear is more viable for implantation
  - Inner ear malformations are present in approximately 20% of children with congenital hearing loss (Vincenti et al, 2014)

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Cognition and Language Skills
  - May be difficult to assess in children with severe developmental delays or very young children as it may be subjective in nature
  - Important to consider the child's communicative intent
  - "Pseudo Handicap Effect" – coexisting disabilities increases the overall disability (Corrales and Oghali, 2014)

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Quality of Life Considerations
  - The World Health Organization defined Quality of Life is defined as the individual's perception of their position in life, in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. (Edwards, Hill and Mahon, 2012)

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Quality of Life Considerations
  - If the child experiences a greater awareness of his surroundings and improves his ability to communicate his desires or needs, the cochlear implant has a positive impact on Quality of Life

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

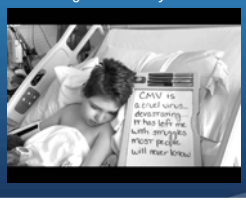
- Quality of Life Considerations
  - Historically, cochlear implants are utilized with the intent of improving hearing for learning auditory based language
  - This may still be achieved but with some children who have additional disabilities, poorer progress is typically observed on traditional measures of speech perception or language acquisition.

Children with Multiple Medical Diagnoses: Cochlear Implant Evaluation Considerations

- Quality of Life Considerations
  - Cultivating family support and appropriate expectations and motivation
    - Enroll the child and family with a language therapist prior to implantation
    - Monthly hearing aid checks with the CI team audiologist to promote the opportunity for discussion

Medical Diagnoses Associated with Hearing Loss

- Congenital Cytomegalovirus (CMV) Infection
  - Most frequent infectious cause of hearing loss
  - 90% of affected patients do not show typical symptoms
  - Associated disorders may include autism, learning disabilities or cognitive delays



Medical Diagnoses Associated with Hearing Loss

Cytomegalovirus in infants

- jaundice (yellowish coloring of the skin)
- enlarged liver
- enlarged spleen
- petechiae (skin rash resulting from bleeding in the skin)
- pneumonia
- central nervous system damage with small head size, brain abnormalities, eye problems or hearing loss

Medical Diagnoses Associated with Hearing Loss

- Usher Syndrome
  - Most common autosomal recessive (AR) cause of hearing loss
  - Associated with vestibular dysfunction
  - Progressive loss of vision
  - Early implantation prior to visual impairment is recommended

Medical Diagnoses Associated with Hearing Loss

- CHARGE Syndrome
  - Rare congenital disorder – more than 90% of children with CHARGE have hearing loss
  - CHARGE stands for: Coloboma, Heart Defects, Atresia of the choanae, Retardation of growth and development, Genital/urinary abnormalities, Ear abnormalities and/or hearing loss

Medical Diagnoses Associated with Hearing Loss

- CHARGE Syndrome
  - Nearly all children with CHARGE have Semicircular Canal Aplasia, Cochlear Nerve Deficiency and Vestibular Dysplasia
  - Many have incompletely developed cochleas
  - Surgical considerations include atypical anatomic landmarks, abberant facial nerve course, abnormal cochlear anatomy
  - CI should be considered due to potential for visual impairment
  - Quality of Life improvements tend to be more related to awareness of sound as speech understanding is not often achieved in this population

Medical Diagnoses Associated with Hearing Loss

- Down Syndrome
  - Low muscle tone
  - Flat facial features, small nose, upward slanting eyes
  - Small, low set ears
  - Hyperflexibility
  - 50% will have cardiac defects
  - 10% of children with Down Syndrome will have Thyroid disease
  - 50%+ have vision issues
  - 10-12% will have gastrointestinal disorders requiring surgical intervention

Medical Diagnoses Associated with Hearing Loss

- Down Syndrome
  - Recurrent otitis media
  - Cognitive delays
  - Hypoplastic cochleas
  - Otitis media management is critical due to increased risk of post operative meningitis – PE tube placement may be necessary
  - Due to range of cognitive delays, the rehabilitation process may be longer with this population

Medical Diagnoses Associated with Hearing Loss

- Cerebral Palsy
  - May range from very mild to severe
  - Low muscle tone/floppy
  - Muscle spasms
  - Poor muscle control, reflexes and posture
  - Delayed physical development
  - Feeding difficulties

Medical Diagnoses Associated with Hearing Loss

- Cerebral Palsy
  - May show difficulties with:
    - Swallowing
    - Breathing
    - Head or neck control
    - Bladder or bowel control
    - Feeding
    - Dental issues
    - Digestive issues

Medical Diagnoses Associated with Hearing Loss

- Cerebral Palsy
  - Communication may be impacted by oral motor weakness
  - 10% have severe vision impairment
  - 50% have a cognitive delay
  - 1 in 50 will have profound hearing loss
  - Hearing loss may be diagnosed later due to the severity of other medical diagnoses
  - 35% of affected individuals are non ambulatory, therefore a special consideration is the positioning of the sound processor microphone and retention of the headpiece

Medical Diagnoses Associated with Hearing Loss

- Cleft Lip/Palate
- Affects up to 7,000 babies in the US annually
  - Cleft lip – split or opening in the lip, which may be large enough to connect the upper lip and nose
  - Cleft Palate – incomplete closure of the roof of the mouth.
    - Hard Palate – bony front portion
    - Soft Palate – soft tissue in the back of the mouth

Medical Diagnoses Associated with Hearing Loss

- Cleft Lip/Palate
  - Frequent episodes of otitis media
  - Speech/articulation issues
  - Feeding difficulties
  - Dental problems
  - Socio-emotional issues due to differences in appearance
  - Surgical repair is recommended within the first year of life

Medical Diagnoses Associated with Hearing Loss

- Cleft Lip/Palate (Galani et al, 2011)
  - 89% of patients experience conductive hearing loss
  - 8% experience mixed hearing loss
  - 3% experience sensorineural hearing loss
  - Medical management of chronic conductive component is critical in this population

Medical Diagnoses Associated with Hearing Loss

- Waardenburg Syndrome
  - Autosomal Dominant
  - Sensorineural Hearing Loss
  - Iris pigmentary abnormality – two different color eyes
  - White forelock
  - Wide set eyes, broad nasal root
  - Low hairline
  - Intestinal defects
  - Spinal defects

Medical Diagnoses Associated with Hearing Loss

- Waardenburg Syndrome
  - Type I and II are most common forms. Type III and IV are rare
  - Accounts for 2% of congenital profound hearing losses

Medical Diagnoses Associated with Hearing Loss

- Autism Spectrum Disorder
  - Extreme unresponsiveness to others
  - Communication deficits
  - Poor turn taking skills
  - Difficulty managing emotions
  - Rigid and repetitive behaviors
  - 1 in 68 children have ASD
  - Greater incidence in boys

Medical Diagnoses Associated with Hearing Loss

- Autism Spectrum Disorder
  - Approximately 3-4% have profound hearing loss
  - Delayed developmental milestones
    - Poor eye contact
    - Resistant to touch, cuddling or hugging
    - Does not engage in typical play with toys

Medical Diagnoses Associated with Hearing Loss

- Autism Spectrum Disorder (Leach and LeBeau, 2014)
  - Sensory integration issues require special consideration
    - Tolerance/consistent use of hearing devices
    - Challenges with making earmolds or keeping hearing aids in place
    - Dislike of light pressure
    - Demonstrates challenges with loud sounds or specific sounds
    - Overall ability to transition or difficulty with change
    - Communicative Intent

Medical Diagnoses Associated with Hearing Loss

Congenital Rubella

- 58% of infants will have profound sensorineural hearing loss
- Vision problems
- Cardiac problems

Medical Diagnoses Associated with Hearing Loss

Individuals with Congenital Rubella may also display:

- Developmental Delay
- Autism Spectrum Disorder
- Schizophrenia
- Growth Retardation
- Learning Disabilities
- Diabetes
- Glaucoma

Medical Diagnoses Associated with Hearing Loss

Donnai Barrow Syndrome – Autosomal Recessive inherited disorder

RARE – only a handful of cases reported

- Unusual facial features: wide set eyes, short nose and flat nasal bridge, posteriorly rotated ears, widows peak hairline
- Sensorineural hearing loss
- Extreme nearsightedness
- Underdeveloped or absent corpus callosum
- Diaphragmatic Hernia
- Omphalocele
- Mild to moderate cognitive delays

Concluding Remarks

- Children with multiple medical diagnoses are not precluded from cochlear implantation
- Family should be actively involved in the candidacy process
- Consideration should be given to improvements in Quality of Life vs traditional anticipated benefits of auditory based language learning
- Family support and expectations/motivation are key to success

References

- Leach, M and LeBeau, V. Deafness with Autism: A School Age Communication Perspective. Presented on Audiology Online, November, 2014.
- Corrales, C and Oghali, J. Cochlear Implant Considerations in Children with Additional Disabilities. Curr Otorhinolaryngol Rep, 2013 June 1(2): 61-68.
- Hang, A, Kim, G and Zdanski, C. Cochlear Implantation in Unique Pediatric Populations. Curr Opin Otolaryngol Head Neck Surg, 2012 December; 20(6) 507-517.

References

- Wakil, N et al. Long Term Outcome After Cochlear Implantation in Children with Additional Developmental Disabilities. International Journal of Audiology 2014; 53: 587-594.
- Edwards, L, Hill, T and Mahon, M. Quality of Life in Children and Adolescents with Cochlear Implants and Additional Needs. International Journal of Pediatric Otorhinolaryngology, 2012 (76) 851-857.
- Gani, B, Kinshuck, A and Sharma, R. A Review of Hearing Loss in Cleft Palate Patients. International Journal of Otolaryngology, vol 2012, article ID 548698.

References

- Palmieri, M et al. Evaluating Benefits of Cochlear Implantation in Deaf Children with Additional Disabilities. Ear and Hearing, 2012, vol 33(6), 721-730
- Rafferty, A et al. Cochlear Implantation in Children with Complex Needs-outcomes. Cochlear Implants International, 2013, vol 14(2), 61-66.
- Vincenti, V et al. Pediatric Cochlear Implantation: An Update. Italian Journal of Pediatrics, 2014, 40: 72, 1-7.

References

Cosetti, M and Waltzman, S. Outcomes in Cochlear Implantation: Variables Affecting Performance in Adults and Children. Otolaryngol Clin N Amer. 2012 Feb; 45(1):155-171.